Modern Concepts of Cardiovascular Disease

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THE DIAGNOSIS AND TREATMENT OF CARDIOVASCULAR SYPHILIS

Part I

The field of cardiovascular syphilis is so broad that no attempt will be made to cover it all in this paper. Instead some of its less well known aspects will be emphasized, newer knowledge presented and a practical guide for treatment outlined.

In order to understand certain of the clinical aspects of cardiovascular syphilis it is necessary to be familiar with a few facts in the pathogenesis of the condition. Many of us think of it as a late sequel of an infection acquired many years earlier. It is incorrect to say that syphilitic aortitis and aortic insufficiency develop on an average twenty years after the primary infection. It is true that these conditions are usually first diagnosed then but they have been developing slowly ever since the time of the chancre. The spirochetes enter the blood stream and invade many of the tissues of the body while the chancre is still present and during the period of the secondary rash. Many of them are destroyed in the lungs but some enter the lymph spaces between the alveoli and pass to the lymph nodes at the hila of the lungs. These are in close relation with the arch of the aorta. From these structures they are thought[1] to pass by retrograde lymph flow to the perivascular lymph spaces about the vasa vasorum in the adventitia of the aorta. Slowly they begin their work of injury to the media and intima of this vessel. Syphilitic aortitis begins immediately, not twenty years after infection. The methods of diagnosis at our disposal do not make it possible for us to detect its presence as soon as the aorta has been invaded but if one keeps looking for it each year after infection it will be discovered much sooner than has been done in the past.

Syphilitic aortitis is the basic pathologic lesion from which all but one of the other manifestations of cardiovascular syphilis develop. Aneurysms appear as the result of weakening and out-pouching of a localized area of the wall of the aorta. Syphilitic aortic insufficiency results from aortitis at the base of the aortic valves. The aortic ring widens, the commissures separate and the valves become incompetent. The syphilitic process extends to the edges of the valve leaflets by way of small vessels in the commissures. Syphilis of the coronary arteries is limited to the mouths of these vessels as they penetrate the aortic wall in the sinuses of Valsalva. A patch of aortitis encroaches upon the mouth of one or both of the coronary arteries and slowly narrows them, sometimes to the point of complete obliteration. The process is very slow and collateral circulation develops so that cardiac infarction seldom if ever results from this lesion. Occasionally the mouths of both coronary arteries are found occluded. The syphilitic process only affects the first few millimeters of these vessels as they penetrate the aortic wall. Lesions beyond this point are attributable usually to arteriosclerosis. Syphilitic myocarditis is such a rare lesion that its existence is still in doubt but gummata of the myocardium occur occasionally. They are one of the causes of sudden and unexpected death and are therefore most numerous in the experience of the Medical Examiner's office.

Since uncomplicated aortitis is the basic lesion of cardiovascular syphilis and since aneurysm, aortic insufficiency and syphilitic coronary osteal stenosis are late sequellae, it is clearly desirable to be able to diagnose this condition before these serious complications have appeared. Authorities differ as to the possibility of doing this. In 1932 Moore[2] studied the clinical records of 105 cases of uncomplicated syphilitic aortitis which had been discovered at necropsy at the Johns Hopkins Hospital between the years 1910 and 1930. Only four had been diagnosed ante mortem and in only thirteen others had the diagnosis been suspected. In 1937 White and Wise[3] stated: "Since a clinical diagnosis of aortic syphilis can rarely be made within ten years after infection, it may safely be said that the early diagnosis of cardiovascular syphilis is practically impossible. If this is so, our interest is naturally next concentrated on the problem of the earliest possible diagnosis of such disease." We have been interested in this problem in the Brooklyn Hospital Cardiac Clinic for the past fourteen years. During the first eight years of the study the diagnosis of uncomplicated aortitis was made in 59 (17.05 per cent) of the 346 syphilitic patients then in the series. The diagnosis was attempted in patients up to 50 years of age. Now after fourteen years experience we have become more cautious and stricter in our criteria. The age limit has been reduced to 40 years. As a result there are only 12 cases (2.5 per cent) of uncomplicated aortitis among the 477 patients now included in the study.

In view of this what are the rules to guide the physician in arriving at the diagnosis of uncomplicated aortitis? First, he must not wait for the patient to come to him complaining of symptoms referable to the heart. Uncomplicated syphilitic aortitis is a symptomless disease. The retrosternal burning pain sometimes ascribed to it is rarely encountered and is probably the result of syphilitic coronary osteal stenosis. Second, he must suspect the possibility of the presence of aortitis in every patient infected with syphilis and examine him regularly at least once a year with this in mind. The most useful method at his disposal is the roentgen ray. Fluoroscopy, orthodiography and teleroentgenography should all be employed whenever possible. If a choice of these methods has to be made, fluoroscopy should be selected. The size and shape of the vascular pedicle should be studied. If its right border extends as far to the right as the margin of the right auricle, the vascular pedicle

should be considered widened. The examiner then studies the contour of the aorta to discover bulging or abnormal pulsation either generalized or localized. In doing this the right and left oblique views are especially valuable. Elongation and increased tortuosity of the aorta can be observed and calcified plaques in the aortic knob can be visualized. Their presence makes the diagnosis of arteriosclerosis of the aorta certain. The size and shape of the heart should be carefully noted. In uncomplicated aortitis the heart is not enlarged. If it is aortic insufficiency, syphilitic coronary disease or some other form of heart disease that produces strain on the left ventricle is responsible for enlargement. An orthodiagraphic tracing can be made during the fluoroscopic examination and kept for purposes of record. If this is not possible then a teleroentgenogram should be taken. It is important to emphasize here that it is impossible to make a diagnosis of aortitis on the basis of X-Ray studies alone. The roentgenologist should describe his findings and state whether or not in his opinion the vascular pedicle shows widening or abnormalities in contour and whether or not the heart is enlarged. Further than this he cannot safely go.

There are no physical signs that are pathognomonic of uncomplicated syphilitic aortitis. A tambour like quality to the aortic second sound has been described as characteristic of this disease. In our own clinic, study of this point in a large number of patients has not enabled us to distinguish between the aortic second sound heard in arteriosclerosis, hypertension or syphilitic aortitis. One clinical point of some value has been derived from this attempt. If a hollow, accentuated aortic second sound is heard in a patient who is under forty years of age and who does not have hypertension, syphilitic aortitis should be carefully considered as the possible cause of the change in quality of this sound. The electrocardiogram is not helpful in the diagnosis of uncomplicated aortitis. Any significant changes in the curves which are found result either from syphilis or arteriosclerosis of the coronary arteries, gumma or aortic insufficiency.

As the result of recent work[4] in the study of uncomplicated syphilitic aortitis it is possible to derive six criteria for the diagnosis of this condition.

1. The patient must be 40 years of age or younger. Above this age arteriosclerosis of the aorta appears to a significant degree. Ophüls in a statistical survey of 3,000 autopsies found that between ages 30 and 40 arteriosclerosis was slight and appeared in only 6 per cent of the cases wheras between 40 and 50 nearly one fourth of all cases showed vascular lesions. It is therefore impossible for the clinician to say whether syphilis or arteriosclerosis or a combination of both is responsible for widening of the aorta apearing after the age of forty.

2. The patient must have had syphilis beyond reasonable doubt. The history of a chancre plus a strongly positive Wassermann Test, precipitin reaction or dark field examination is satisfactory evidence. If there is no history of infection, then two strongly positive standard serological tests are necessary to establish the diagnosis of syphilis. If neither of these conditions is satisfied then there must be incontrovertible evidence of syphilis elsewhere in the body; tabes dorsalis or general paresis for example. If one attempts to diagnose syphilitic aortitis in patients in whom the presence of syphilis is doubtful many errors will be made.

3. There must be no evidence of any other disease that might dilate the aorta. Arteriosclerosis, rheumatic fever, rheumatic heart disease, hypertension and hyperthyroidism must be excluded.

4. There must be roentgenographic or fluoroscopic evidence of a dilated aorta.

5. There may be a hollow, accentuated aortic second sound.

6. There may be a systolic murmur at the aortic area.

These criteria have served very well in diagnosing uncomplicated aortitis in patients 40 years of age or younger. They can be used equally well in patients over 40 but in this group the diagnosis should be recorded: syphilitic aortitis or arteriosclerosis of the aorta. If evidence of a dilated aorta is discovered in a patient over 40 it is impossible for the physician to say, unless aneurysm or aortic insufficiency is present, whether syphilis or arteriosclerosis or a combination of both conditions have caused the dilatation. Further these two diseases are found together very often at post mortem examination. Therefore both on clinical and pathologic grounds the diagnosis syphilitic aortitis or arteriosclerosis of the aorta is sound. In our study of 477 syphilitic patients there are 86 patients (18 per cent) in whom this diagnosis has been made. Among them 5 have developed aortic insufficiency and 1 an aneurysm while under observation.

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